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Case Report

A challenging case of a tanycytic ependymoma – A case report

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ABSTRACT

Intramedullary spinal cord neoplasms are among one of the rare tumors and account for 4% to 10% of all the central nervous system tumors. Spinal cord ependymomas are the most common type of tumors in adults, and astrocytomas of the cord are most common in children. Among the ependymomas, Tanycytic type is a rare one that usually arises in the intramedullary part of the spine. The histology is unique which is emphasized by the absence of ependymal pattern of cells but has close resemblance to schwannoma and astrocytoma which poses a diagnostic challenge. Hereby we present a case of a young male diagnosed as tanycytic ependymoma but was initially thought to be a glioma clinically. The histology showed spindle cells with oval and elongated nuclei in fascicles in a fibrillary background suggesting a preliminary diagnosis of glioma / schwannoma. However with the help of Immunohistochemical staining, a strong immunoreactivity for glial fibrillary acidic protein (GFAP) with S-100 positivity, confirmed that the tumor was a tanycytic ependymoma. This case underlines the importance of accurate diagnosis of CNS tumors with the help of age, location, detailed histopathological examination and immunohistochemistry to differentiate from other neoplasms as treatment differs for different tumors.

1. Introduction

Tumors in the spine comprise about 15% of all tumors in the central nervous system. They usually are benign and cause symptoms primarily through compression of the spinal cord and nerves. Spinal tumors can be classified into three groups, based on their location: extradural, intradural-extramedullary, and intramedullary. Extradural tumors are most common, as they occupy the vertebrae body or structures outside the dura. They are most commonly metastatic. Intradural extramedullary tumors are the second most common and come from the leptomeninges or nerve roots. These tumors are located inside the dura but external from the spinal cord, exemplified by meningiomas or neurofibromas. The least common (2 to 5%) are intramedullary spinal cord tumors, these arise from the spinal cord proper, leading to invasion and destruction of the gray and white matter. Ependymomas and astrocytomas are the most commonly encountered intramedullary spinal cord tumors, followed by hemangioblastomas.[1] About 80% of intramedullary tumors are gliomas, which can be subdivided into astrocytomas and ependymomas. Astrocytomas are more common in children, while ependymomas are more often found in adults with intramedullary spinal cord tumor. Astrocytomas peak in the third to fifth decades, often low-grade, and are found most commonly at the thoracic level. Ependymomas are more commonly found in the lower cord, conus, and filum, with a slight male predominance and peak in the third to sixth decades. A rare form of ependymoma, tanycytic type are derived from tanycytes which are elongated spindly bipolar cells, and hence are misdiagnosed as a nervous system

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tumor of the spindle cells. Definitive diagnosis requires pathological analysis, including histological characteristics and immunohistochemistry.

2. Case History

A 34 year old male presented with history of low back pain for 3 months, history of neck pain radiating to hands and burning sensation of foot since 2 weeks. On examination the patient was conscious and oriented, had tenderness over upper dorsal region. On MRI, an intramedullary space occupying lesion extending from C6 to D1 was seen. Radiologically a diagnosis of glioma was given.

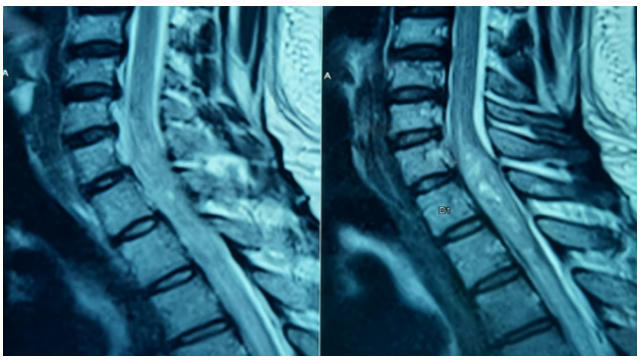

Fig. 1: Intramedullary space occupying lesion occupying C6 to D1 was seen on MRI.

An elective procedure laminectomy was done and the specimen was sent for histopathology. Histopathology department received multiple grey white soft tissue altogether measuring 1.5 cc in aggregate. All the tissue was processed.

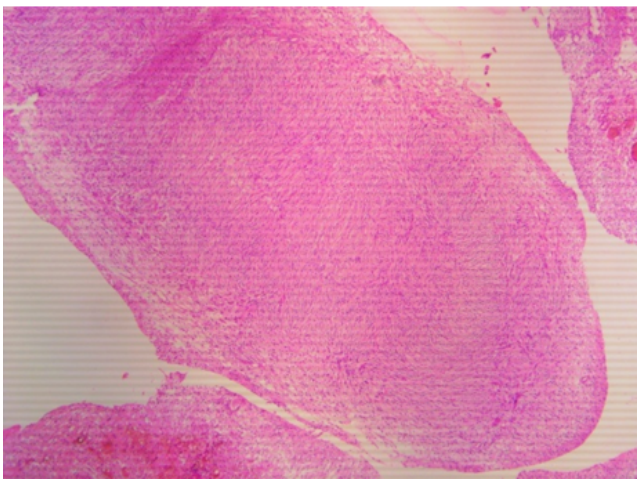

Fig. 2: Lobular architecture of the tumor on low power (H&E 10X)

On histology, a lobular cellular neoplasm (Figure 2) composed of oval to spindle cells in whorls, interlacing bundles (Figure 3) in a prominent fibrillary background

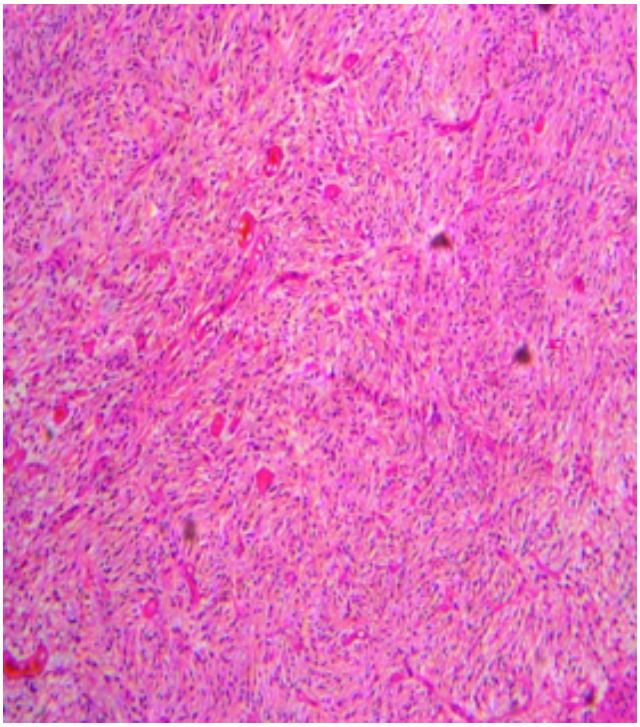

Fig. 3: Spindle shaped cells in intersecting fascicles (H&E 20X)

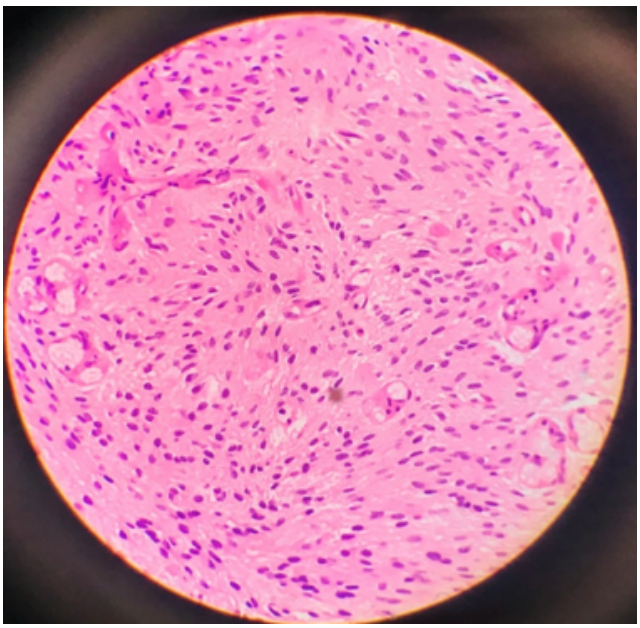

Fig. 4: High power view displaying bland elongated cells in fibrillary background with interspersed blood vessels (H&E 40X)

(Figure 4) was seen. Moderate pleomorphism of cells with occasional giant cell and moderate degree of mitosis 1 to 2 / high power field was seen. The lesion was highly vascular with foci of hemorrhage and delicate vascular channels traversing the parenchyma. A differential diagnosis of schwannoma and pilocytic astrocytoma was considered. On immunohistochemistry: GFAP, S100, ATRX and vimentin was positive. Other markers like OLIG2 and p53 was patchy positive. EMA was negative and Ki 67 proliferative index was 10 to 12%. The patient was not given any adjuvant radio / chemotherapy. At 6 months follow up, the patient has improved and is ambulatory.

3. Discussion

4% to 10% of all central nervous system tumors are comprised by intramedullary spinal cord neoplasms which are rare in occurrence. Among adults, Spinal cord ependymomas are the most common type whereas in children the common type is astrocytomas. 70% of all intramedullary neoplasms is constituted by both these tumors.[2] Patients with intramedullary spinal cord tumors most commonly present with pain, which typically worsens at night when the patient is non ambulatory and pain can be radicular type or diffuse in nature. If dermatomal symptomology does not correlate clinically with disc herniation, intramedullary spinal cord tumors should be suspected. The pain can be burning type, local or bilateral, causing stiff neck or back. The tumor can compress the motor or sensory nerves leading to changes such as paresthesia, followed by motor disturbances. Intramedullary tumors are sporadic in most of the cases, but some have association with clinical syndromes such as Von Hippel-Lindau disease (VHL) and neurofibromatosis 1, 2 (NF-1, NF-2).

Ependymoma is a type of glioma which is circumscribed and comprised of uniform sized small cells having round nuclei in a fibrillary matrix and is characterized by perivascular anucleate zone. Ependymal rosettes is seen only in around one fourth of cases.[3]

Classic type of ependymomas mainly occur intracranially, they also occur in the spinal cord but myxopapillary variant of ependymoma is more common in that site. Spinal ependymoma have late recurrences (>5 years after surgery). They have a significantly better outcome than the intracranial ependymomas. WHO grade II and III correspond to classic ependymoma and anaplastic ependymoma respectively. However, there is no established association between grade and biological behaviour or survival.

Friede and Pollak, first described the tanycytic variant of ependymomas in detail as having an appearance similar to tanycytes (ependymoglia) which is a common ancestor of both ependymal cells and astrocytes.[4]Tanycyte cells are considered to be the origin in two types of CNS tumors, that is - tanycytic ependymoma and astroblastoma. As a result of this derivation, tanycytic variant of ependymomas were presumed to be found in the spinal cord typically where the raphe are abundant in tanycytes and ependymoglia.

Under light microscopy, Tanycytic ependymoma cells are similar to other tumor cells with similar features, leading to a challenging diagnosis. The tanycytic ependymoma are low to moderately cellular and is composed of bipolar cells predominantly with long processes which has a spindled to piloid appearance. These characteristic features can mimic other tumor types.[5]

Pilocytic astrocytoma is considered a differential due to the piloid appearance of the bipolar cells with particularly long processes. Some of the distinguishing features that set the tanycytic ependymoma cells apart from astrocytoma are the presence of large ovoid nuclei, tight perivascular packing of cells, absence of Rosenthal fibers and their isomorphic cellular appearance.[6]

Per operatively frozen section diagnostic uncertainty occur due to the presence of spindled type cells which mimics schwannomas.[7] One distinguishing feature is that tanycytic ependymoma cells have more uniform oval nuclei as compared to those of schwannoma cells. In these cases, final definitive diagnosis is aided by immunohistochemical staining. Tanycytic ependymomas tend to have diffuse positive staining for vimentin, GFAP and variable S -100 staining, whereas schwannomas commonly have strong, diffuse S -100 staining, and are typically negative for GFAP. Similar to other ependymal neoplasms, tanycytic variant shows ring and dot like perinuclear staining for EMA which represent intracytoplasmic microrosettes.

On radiology, after administration of contrast agent ependymoma enhances on T1- weighted imaging. It is often associated with a syrinx or hematoma. Ependymoma almost always occurs in intramedullary location whereas schwannoma almost always occurs in extramedullary location which also enhances on T1-weighted imaging with gadolinium contrast enhancement. The other differential diagnosis, Pilocytic astrocytomas rarely enhance with contrast.

Currently, complete surgical resection of the tumor is the primary treatment method for Tanycytic ependymoma. For patients undergoing successful complete surgical resection, postoperative radiotherapy is not considered necessary. For patients with postoperative recurrence, surgical resection with postoperative radiotherapy is the modality of treatment considered.

4. Conclusion

This case illustrates tanycytic ependymoma which is an unusual variant of ependymal neoplasm, This variant is also known as fibrillary ependymoma, presenting as an intramedullary mass that is difficult to distinguish from a schwannoma / pilocytic astrocytoma on frozen section or

on histological examination. The tanycytic ependymomas typically doesn't have the classic histology of ependymal rosettes or perivascular pseudorosettes which makes the diagnosis a challenge for the pathologists. In our patient, a correct diagnosis was only reached with the support of immunohistochemical stains. This entity should be considered in the differential diagnosis of gliomas as they have better prognosis and do not require radiotherapy.

5. Conflict of Interest

The authors declare no relevant conflicts of interest.

6. Source of Funding

None.

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